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Adenocarcinoma in a Duplicated Bladder

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We report a rare case of duplication of the bladder, urethra, uterus, vagina, and associated anomalies in a woman. As an infant, she initially underwent successful surgical reconstruction. As an adult, she developed adenocarcinoma within the defunctionalized bladder moiety. The surgical management and pathology of this cancer are detailed and the pertinent literature reviewed. *J. Surg. Oncol. 1997;65:280–283.* © 1997 Wiley-Liss, Inc.

KEY WORDS: carcinoma; bladder duplication; pathology; embryology

INTRODUCTION

Complete duplication of the bladder is rare, with <50 cases previously reported in the world literature [1–13]. We report here the first case of adenocarcinoma arising in a duplicated bladder. We also detail our surgical management and review the literature.

CASE REPORT

A white female, 58 years old at the time of this report, was a product of a normal gestation and delivery. She was born with bifid external genitalia, as well as complete duplication of the urethra, bladder, vagina, and uterus. Further investigation revealed ventricular and atrial septal defects and a normal solitary kidney on the right, which drained into a right hemibladder, with its own urethra. The left hemibladder was defunctionalized, yet also had a separate urethra. As an infant, she underwent vaginal reconstruction, transabdominal hysterectomy, resection of a diverticulum of the right hemibladder, and repair of her cardiac anomalies.

The patient remained in relative good health until 57 years later when she developed gross blood per urethra. A renal ultrasound and intravenous urogram showed an enlarged, otherwise normal-appearing solitary kidney, which drained into an ipsilateral hemibladder. Cystoscopy of the right urethra noted a bladder with a hemitrigone, a single ureteral orifice (normal in size and shape), and normal urothelium. Cystoscopy of the left urethra and of the defunctionalized hemibladder also noted a hemitrigone and single ureteral orifice. However, this bladder was filled with copious necrotic debris, as well as a 2 cm papillary-appearing tumor located along the left medial wall. Urine cytology revealed atypical glandular cells suggestive of adenocarcinoma. The tumor was subsequently resected transurethrally and the bladder randomly biopsied. Pathologic examination demonstrated a localized low-grade adenocarcinoma with invasion of the lamina propria.

Metastatic workup was negative. Chest radiograph, barium enema, and mammography were also unremarkable. Computed tomography (CT) noted no extravesical tumor extension or retroperitoneal adenopathy. Magnetic resonance imaging (MRI) of the pelvis showed a duplicated bladder that shared a common wall medially, with a nodular tumor mass along this common wall (Fig 1.).

The patient underwent a left hemicystectomy, vaginoplasty, left pelvic lymph node dissection, and right hemicystoplasty via a midline abdominal incision. Initially, foley catheters were placed in each bladder, with the left filled with methylene blue and saline and the right only saline. Upon opening the abdomen, the urachus was dissected down to the dome of the left hemibladder, the left bladder pedicle taken down sharply, and the right hemibladder opened. To protect the right ureter, a 6 Fr pediatric feeding tube was inserted in the ureteral orifice. The entire left hemibladder, the bladder common wall, and the ipsilateral urethra were then resected en bloc. In order not to render the patient incontinent, great care was taken during urethral dissection not to damage the contralateral

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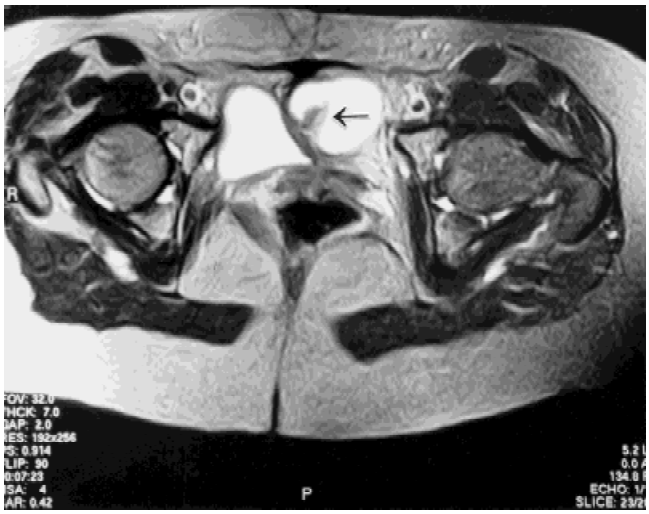

Fig. 1. T2 weighted axial sequenced magnetic resonance image demonstrating the duplicated bladder and shared common wall. Note the nodular mass along the medial wall of the left hemibladder, which represents the primary bladder cancer (see arrow).

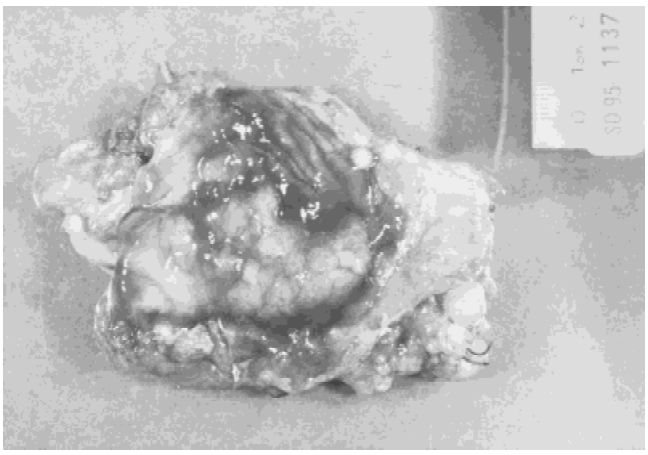

Fig. 2. Gross pathologic examination of the open hemi-bladder, showing the polypoid tumor mass on the medial wall.

urethral sphincter. The right hemibladder was reconstructed by reapproximating the edges in two layers and placed at rest with a foley catheter. Estimated bladder volume was 150 cc, and augmentation cystoplasty was not considered at this time. A pelvic foley drain was left in the prior left urethral opening.

Gross pathologic examination revealed a 6.5 × 3.5 × 2.5 cm left hemibladder, a 1.4 cm ipsilateral urethra, and a 4.5 × 2.0 cm light-tan mucoid mass along the medial bladder wall (Fig. 2). The bladder surface was edematous, light tan, and covered with thick red-brown, mucoid material throughout. On sectioning, there was no gross tumor invasion of the bladder wall. Microscopic examination demonstrated a low-grade papillary mucinous adenocarcinoma, with no evidence of muscularis propria invasion (Fig 3A,B). The mucosa away from the tumor was columnar with various grades of dysplasia/carcinoma in situ. All margins of resection were negative for tumor, and all four pelvic lymph nodes were negative for metastatic disease.

After an uneventful postoperative course, the patient was discharged home on day 5. Two weeks later a cystogram showed no extravasation, and the foley catheter was then removed. During the following 2 years, the patient was followed at appropriate intervals and has shown no evidence of disease based upon endoscopic, cytologic, and CT evaluations. She has also continued to maintain good urinary continence and control, but experiences urinary frequency every 4 hours.

DISCUSSION

Bladder duplication can be complete or incomplete. With incomplete duplication, the two bladder units communicate and drain into a single urethra and usually have no associated genital or anorectal malformations [2]. In complete duplication, there are two bladder halves, each half with its own ipsilateral ureter and each draining into separate urethras. In nearly all cases (90%), the genitalia are also duplicated [2]. In women, there are usually two vaginas (89%) and a duplicated uterus, ranging from a bicornuate uterus to two uteri, each with a single uterine horn and oviduct. In men, there are usually either two penises, one partially duplicated penis, or one penis with two distinct urethras. The ventral urethra is usually the functional one. The scrotum is commonly bifid. There are also reports of cryptorchid testes and hypospadias. Associated anomalies with complete bladder duplication are myriad, and include: (1) colon duplication and other anorectal malformations, e.g., anal stenosis, ectopic, and imperforate anus (40–56%), (2) fistulas between the rectum vagina and urethra, (3) renal ectopia, (4) ileal duplication (usually from Meckel's diverticulum till the ileal-cecal valve), (5) spinal anomalies—sacral and coccygeal vertebral column duplication, meningocele, and myelomeningocele (10–15%), (6) other skeletal anomalies (e.g., symphysis pubis diastasis,), (7) cardiac anomalies (ventricular septal defects, atrial septal defects), and (8) gut malrotation and intussusception. Of the above urologic anomalies, most are relatively asymptomatic and do not require surgical reconstruction. However, patients who are incontinent, who have recurrent urinary infections, lower or upper urinary tract obstruction, or voiding dysfunction such as double urinary streams usually merit urologic repair [1–13].

The embryogenesis of bladder duplication is unclear. It has been hypothesized that bladder duplication was secondary to a membrane that initially obstructs one of the ureters at the vesical junction, which then dilates [2]. The primitive bladder then develops around this dilated ureter, resulting in a two-part bladder. Ravitch [3] theo-

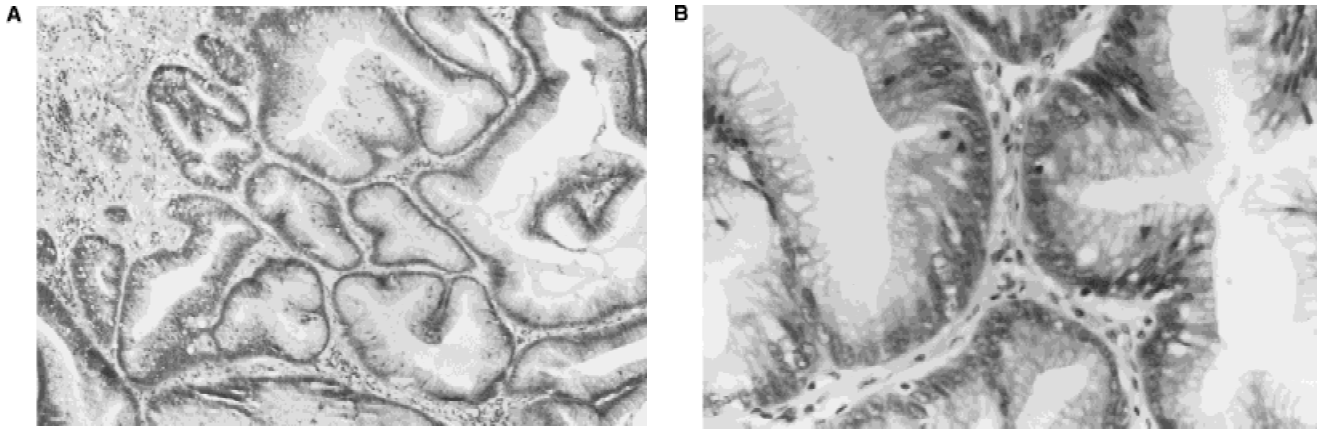

Fig. 3. **A.** Histologic section of the bladder tumor demonstrating a glandular, papillary pattern. H & E, reduced from original magnification ×100. **B.** Higher power, demonstrating moderately pleomorphic columnar cells with a papillary formation. H & E, reduced from original magnification ×210.

rized that the etiology was twinning of the embryo tail, which also would explain the often associated bowel (cloacal) duplication. However, this does not explain the relative rarity of skeletal duplication, which in theory should be more common. Satter and Mossman [4] hypothesized the presence of a ''double allantoic duct'' where a persistent urogenital (UG) sagital septum divides the endoderm into two halves and thus forms two bladders, urachuses, and UG sinuses. The two UG sinuses then cause the urethral plate to widen, which on canalization forms into two separate urethras. Again, this theory cannot explain the associated skeletal anomalies. Overall, there is no consensus or clear support for either of the above theories.

Adenocarcinoma of the bladder is a rare cancer representing only 0.5–2.0% of all bladder cancer [14]. In origin, bladder adenocarcinomas are either primary urachal, primary vesical, or extravesical with direct extension or metastases into the bladder. Overall, the majority of bladder adenocarcinomas invade the bladder extravesically, most originating as colo-rectal, prostatic, or uterine primaries. Thus before a definitive diagnosis of primary bladder adenocarcinoma is made, a careful metastatic survey should be performed. Primary vesical cancers usually present with irritative voiding symptoms, suprapubic discomfort, hematuria, as was the case for our patient, and mucinous urine. These tumors are usually solitary and found at the trigone or dome, but can be found in all parts of the bladder. Cystoscopically, the tumor is usually solid but can appear as an ulcerative or papillary lesion, or just as bullous edema. Histologically, these tumors are glandular cancers, with mucin-producing epithelium that cannot be distinguished from colon cancer. Some of these tumors have signet-ring cells, which are particularly aggressive, with mortality rates >50% at 1 year [14]. They are also highly chemoresistant and radioresistant, and radical cystectomy always should be considered.

Primary adenocarcinoma has been associated with bladder extrophy, shistosomiasis, and cyclophosphamide therapy, all presumed secondary to chronic inflammation. We report here the first case of adenocarcinoma arising within a duplicated bladder. We theorize that the etiology of the adenocarcinoma maybe due to the hemibladder being defunctionalized (no ipsilateral kidney), which predisposed the transitional cell lining of the hemibladder to undergo metaplasia. Another possibility is the persistence of an embryonic glandular cell lining to the hemibladder that never underwent transitional cell differentiation. In the future, with defunctionalized bladders, we would advocate periodic bladder screening.

Overall, prognosis for bladder adenocarcinoma is generally poor, with 5-year survival rates at ~20% [14]. Most adenocarcinomas are muscle invasive, poorly differentiated, or metastatic by the time of diagnosis, which helps to explain its relatively poor prognosis. Furthermore, they are resistant to both radiation and chemotherapy. The treatment of choice, then, is surgery. Radical cystectomy with pelvic lymph node dissection has been the preferred management, particularly because after partial cystectomy, local tumor recurrence rates have been very high. Patients with bladder adenocarcinoma who have survived >5 years usually have undergone some form of radical surgery [14].

The goal of surgery on the duplicated bladder and its associated anomalies is the preservation of urinary control and a satisfactory cosmetic result. As was the situation with our patient, management must be tailored to each case. By careful wide en bloc resection of the cancerous bladder, while still preserving the noncancerous duplicated bladder and urethra, we were able to perform

a fundamentally successful cancer operation, as well as avoid the morbidity of incontinence.

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